form should be performed routinely in daily rheumatological prac-
tice including radiological assessment as well as standard evaluation
of peripheral joints. Our data suggest that there is a necessity to
create complex index which includes assessment of tendons/
entheses/peripheral joints for patients with JIA.

**A10.9 EVIDENCE FOR PROGRESSIVE REDUCTION AND LOSS
OF TELOCYTES IN THE DERMAL CELLULAR NETWORK
OF SYSTEMIC SCLEROSIS**

doi:10.1136/annrheumdis-2013-203224.9

1,2Mirko Manetti, 1Serena Guiducci, 1Martina Ruffo, 1Irene Rosa, 1Maria Simonetta
Faussone-Pellegrini, 1Lidia Ibaia-Manneschi, 1Marco Matucci-Cerinic, 1Department
of Anatomy, Histology and Forensic Medicine, University of Florence, Florence, Italy;
2Department of Biomedicine, Division of Rheumatology, AOUC, and Excellence Centre for
Research, Transfer and High Education (EDNDR), The University of Florence, Florence, Italy

**Background and Objectives** Telocytes are a distinct population
of stromal cells which have been recently identified in a wide vari-
ty of tissues and organs, including the skin. By their extremely
long cytoplasmic processes telocytes may act as supporting cells
and form a scaffold to define the correct three-dimensional organ-
isation of tissues/organs during pre-natal life, or their repair/
renewal in post-natal life. Moreover, telocytes may influence the
transcriptional activity of neighbouring stromal cells (fibroblasts/
myofibroblasts, mast cells), either by cell-to-cell contacts or by
secreting paracrine signalling molecules, and may be implicated in
tissue regeneration by cooperating with stem cell niches to form
tandem cell structures. Systemic sclerosis (SSc) is a complex con-
nective tissue disease characterised by fibrosis of the skin and
internal organs. Up to now, most of the studies have focused on
fibroblasts/myofibroblasts, while little is known about the possi-
ble involvement of other stromal cell types in SSc pathophysi-
ology. In the present study, we investigated the distribution and
ultrastructural features of telocytes in the skin of SSc patients
compared with normal skin.

**Methods** Forearm skin biopsies were obtained from 24 SSc patients
(13 limited cutaneous SSc (lcSSc), 11 diffuse cutaneous SSc (dcSSc))
and 10 healthy controls. Skin sections were subjected to immuno-
chemical or immunofluorescence labelling for CD34, CD31/
PECAM-1, alpha-smooth muscle actin (alpha-SMA), CD11c, CD90/
Thy-1, c-kit/CD117 and mast cell tryptase. Ultrathin sections
were processed for transmission electron microscopy (TEM).

**Results** By an integrated immunohistochemical and TEM
approach, we confirmed that telocytes were present in human der-
is, where they were mainly recognisable by their typical ultra-
structural features and were immunophenotypically characterised
by CD34 expression. Dermal telocytes were immunophenotypi-
cally negative for CD31/PECAM-1 (endothelial cells), alpha-SMA
(myofibroblasts, pericytes/vascular smooth muscle cells), CD11c
(dendritic cells/macrophages), CD90/Thy-1 (fibroblasts) and c-kit/CD117
(mast cells). In normal skin, telocytes were organised to
form three-dimensional networks distributed among collagen
bundles and elastic fibres, and surrounded microvessels, nerves and
skin adnexa (hair follicles, sebaceous and sweat glands). Telocytes
displayed severe ultrastructural damages (swollen mitochondria,
cytoplasmic vacuolisation, lipofuscinic bodies) suggestive of isch-
emia-induced cell degeneration and were progressively lost from
the clinically affected skin of SSc patients. Telocyte damage and
loss evolved differently according to lcSSc/dcSSc subsets and
early/advanced stages, being more rapid and severe in dcSSc.

**Conclusions** In SSc skin, the progressive loss of telocytes might
i) contribute to the altered three-dimensional organisation of the
extracellular matrix, ii) reduce the control of fibroblast/myofibro-
blast and mast cell activity, and iii) impair skin regeneration and/or
repair.

**A10.10 EXAMINATION OF IL-6, TNF I AND II RECEPTOR
DISTRIBUTION ON PERIPHERAL BLOOD MONONUCLEAR
CELLS (PBMC) IN SEROPOSITIVE AND SERONEGATIVE
RHEUMATOID ARTHRITIS (RA)**

doi:10.1136/annrheumdis-2013-203224.10

Martyna Skywarak, Babett Haschel, Julia Fantana, Martin Aringer. Division
of Rheumatology, Department of Medicine III, University Clinical Center Carl Gustav Carus
at the Technical University of Dresden, Germany

**Background and Objectives** Despite significant clinical overlap,
genetic evidence and, in part, response to therapy distinguish sero-
positive (RA+), i.e. rheumatoid factor (RF) positive RA patients
from patients with RF and anti-CCP-negative (RA-) disease. The
aim of this study was to profile RA+ and RA- patients with regard
to the differential expression of receptors for IL-6 and TNF, cyto-
kines of established importance in RA.

**Materials and Methods** PBMC of RA+ and RA- patients were
compared to each other and to healthy individuals (HC). Most
(93%) of the RA+ patients were also positive for anti-CCP antibod-
ies. PBMC were immediately prepared from peripheral venous
blood. For determining the percentage of IL-6Ra (CD26), gp130
(CD130), TNFR I (CD120a), and TNFR II (CD120b) positive cells,
PBMC were stained with PE-labelled or control antibodies. Cells
were analysed on a Becton Dickinson FACSCalibur fluorocytome-
ter, gating for lymphocytes.

**Results** Disease duration (median 7.0 (0.04–66) versus 3.5 (0.04–6)
years) and disease activity (CDAI median 15.6 (5.3–54.5) versus
13.5 (4.4–28)) were comparable between RA+ and RA- patients.
Lymphocytes of RA+ and RA- patients differed in their lymphocyte
expression of CD126+ and CD120b+. The percentage of CD126+
lymphocytes in RA+ was decreased in comparison with RA- (mean
± SD, 49 ± 14 versus RA-58 ± 11, p = 0.05) and HC (59 ± 9, p = 0.0007).
The difference between RA- and HC was not signifi-
cant. The percentage of CD130+ lymphocytes in RA+ (51 ± 11)
did not differ to that of RA- and HC (58 ± 11%, p = 0.007). While the mean values for CD130 were similar between RA+ and
RA- (55 ± 14%), the latter values were not significantly different
from HC. In contrast to the IL-6 receptor, CD120b+ lymphocytes
were increased in RA+ patients (69 ± 12% versus RA- 58 ± 12%,
versus HC 57 ± 11, p = 0.0002). Again, the difference
between RA- and HC was not significant. The percentages of
CD120a+ lymphocytes were low in all groups. Nevertheless, mean
percentages of CD120a+ lymphocytes from RA- (1.5 (0.53–2) %
versus HC (median 1 (0.4–4)% p = 0.05) were somewhat higher
than those of RA+ (median 1.1 (0.3–5)%, p = 0.9).

**Conclusions** A direct comparison of (IL-6 mediated) downregu-
lation of CD126 and (TNF mediated) upregulation of CD120b sug-
gests that both are clearly more pronounced in RA+ than in RA-0.
This was not explained by differences in disease activity.

**A10.11 EXPRESSION OF UNFOLDED PROTEIN RESPONSE GENES
IN SYNOVIIUM AND BLOOD MONONUCLEAR CELLS
OF HLA-B27 POSITIVE ANKYLOSING SPONDYLITIS PATIENTS
IS NOT INCREASED COMPARED TO OTHER ARTHRITIS
PATIENTS AND HEALTHY CONTROLS**

doi:10.1136/annrheumdis-2013-203224.11

Barbara Neerinckx, Shea Carter, Rik J Lories. Laboratory of Tissue Homeostasis and
Disease, Skeletal Biology and Engineering Research Center, Department of Development
and Regeneration, KU Leuven, Belgium

**Background** The HLA-B27 heavy chain is prone to misfolding.
Misfolded proteins give rise to endoplasmic reticulum stress and
activation of the Unfolded Protein Response (UPR). The UPR is
strongly activated in HLA-B27 transgenic rats, an animal model for